Epidural metastasis of Ewing’s sarcoma: a rare localisation

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Summary

Epidural spinal metastasis of Ewing’s sarcoma is rarely observed. We report on a rare case of purely epidural spinal metastasis of Ewing’s sarcoma with pain and paraplegia, and describe the treatment and final outcome of the patient.

Key words: epidural, Ewing’s sarcoma, metastasis, radiotherapy

Introduction

Ewing’s sarcoma of the bone is the most common malignant bone tumor in children younger than 10 years of age [1,2]. It is a small round-cell tumor of the bone [3,4]. Metastatic spread is frequent, lungs and bones being the most common sites of metastases [5]. Although Ewing’s sarcoma metastases can involve almost every site, epidural metastasis is only rarely observed. We report on a case of Ewing’s sarcoma metastatic to the epidural space.

Case presentation

A 10-year-old boy had been presented with a mass on the first metatarsal of the left foot with a histologic diagnosis of Ewing’s sarcoma in 1992. He was treated with 14 courses of chemotherapy (doxorubicin 20 mg/m²/day, days 1-3, plus vincristine 1.4 mg/m², day 1, alternating every 3 weeks with actinomycin-D 0.5 mg/m²/day, days 21-23, plus ifosfamide 2 g/m²/day, days 21-23, with mesna (protocol EICESS 92) [6], and he then received 54.4 Gy local radiotherapy. After 4-year disease-free survival, a solitary metastatic mass in the right lung was diagnosed. He was treated with right middle lobectomy and combination chemotherapy with cyclophosphamide 1800 mg/m² plus vincristine 1.4 mg/m² every 3 weeks, alternating with ifosfamide 1.8 g/m²/day, days 1-3 plus mesna, and etoposide 100 mg/m²/day, days 1-5 every 3 weeks for one year. During follow-up, a new metastasis was found in the lower lobe of the right lung in October 2001. Right lower lobe wedge resection was performed and the patient received 6 courses of continuous infusion ifosfamide 2.5 g/m²/day with mesna, days 1-5. He was then referred to our clinic in February 2002. Bilateral lung irradiation up to 18 Gy was delivered and a boost dose of 45 Gy to the metastatic tumor volume was administered. For a long time the patient was without disease-related symptoms. In the 14th month after lung irradiation he presented with back pain persisting for 1 month. Physical and neurological examination, thorax computed tomography, bone scan and biochemistry were all normal. However, 4 days later he developed weakness and numbness in both legs. Neurological examination was unremarkable, except loss of power in the lower extremities (3/5) and hypoesthesia below the cervical (C) 8 level. Magnetic resonance imaging (MRI) of the thoracic
spine revealed an homogenous epidural mass at the thoracic (T) 6- T7 region, relatively hyperintense on T2-weighted images and hypointense on T1-weighted images. The right posterolateral epidural location of the mass compressed the cord to the left anterior portion of the spinal canal. The right foraminal extension of the mass also occupied the periforaminal fatty place (Figures 1 and 2). The MRI study of the brain and lumbar vertebrae was normal. Therefore, a T6-T7 laminectomy and gross total removal of the tumor were performed urgently.

The curettage material of 2 cc volume taken from the extradural T6-T7 space was evaluated histologically. The tumor consisted of undifferentiated primitive small blue round cells (Figure 3). Some areas showed rosetoid configuration of tumor cells. The case was considered as Ewing’s sarcoma metastasis, as the patient had a history of this malignancy. Cytological examination of the cerebrospinal fluid for meningeal involvement revealed no atypical cells but macrophages and lymphocytes.

After spinal decompressive surgery his neurological status partially improved. He could walk and stand up with help. The patient received radiotherapy with 30 Gy in 15 fractions to the T5-T8 region postoperatively. However, his neurological deficit progressed during radiotherapy. Neurological examination revealed rectal and urinary incontinence, hypaesthesia below the level of T6-T7 and positive Romberg sign. The patient was put on dexamethasone 8mg i.v.x3/daily. A new craniospinal MRI was done. The cranial MRI was normal; however, the spinal MRI revealed a pathological signal posterior to the corpus of L1 with an anterior epidural mass. The patient received radiotherapy with 30 Gy in 10 fractions to T12-L2 region. Chemotherapy was planned, but it never started because of the platelet count (59,000/mm³) and his bad general condition. Four days after the radiotherapy started, bilateral abducent nerve paralysis, difficulty in swallowing and loss of hearing developed. Although the cranial MRI was normal, we considered these findings as a cranial infiltration, while he responded clinically to mannitol and dexamethasone treatment. Whole brain irradiation was started and cyclophosphamide 250 mg/m² days 1-3 was administered but 7 days after chemotherapy the patient died due to haemoptysis and febrile neutropenia.
Discussion

Ewing’s sarcoma is an undifferentiated blue round-cell tumor of the bone. It originates from neural tissue, specifically from postganglionic, parasympathetic, primordial cells [3,4]. Seventy-five to 80% of patients with Ewing’s sarcoma present with localized disease at diagnosis. The most common sites of metastases are the lung, bone, bone marrow, soft tissue, brain and spine [5]. According to reports of the Bristol Bone Tumor Register, the initial metastasis in patients with long bone primaries can occur both in pulmonary and extrapulmonary sites. When other bones are involved, there is a tendency for extrapulmonary sites [7]. When the primary tumor is not in the long bones, vertebral metastasis can occur with higher probability due to the Batson’s pathway. Mehda et al. reported central nervous system (CNS) metastasis in 15 out of 27 patients (55.5 %) [8]. However, results from the IESS and the National Cancer Institute studies did not confirm this high incidence of CNS involvement [9,10]. In the Trigg’s et al. review [10] the CNS involvement was 2.2% and in almost all of these patients CNS involvement was associated with metastasis in other sites. Meningeal or intraparenchymal CNS disease was the initial site of recurrence in 3 out of 10 patients.

Extra- or epidural metastases are relatively frequent. However, spinal cord compression with serious complications from malignant diseases in children is rare. Spread pathways of epidural metastases are direct extension of mediastinal or retroperitoneal masses or nodes through the intervertebral foramina or transcotical extension from metastatic disease in the vertebral corpus or haematogenous, paravertebral and epidural plexus venousus [11]. The presenting symptoms are usually sudden pain exacerbation, which is difficult to control with increasing doses of analgesics and worsens with recumbency, cough and Valsalva maneuver, pain with radiculopathy and Lhermitte sign [11]. The onset symptom of our case was only pain, however his pain progressed and paraplegia occurred in just 4 days. The differences in symptoms between epidural and intradural metastases do not differ too much, but they can be distinguished from each other. Bladder and bowel dysfunction is much more present in intradural-extradural metastasis than in epidural metastases. Also, prominent pain which is experienced at an early stage is more severe and crampy and radicular dysfunction is more prominent in intradural-extradural metastasis. Intrametullary metastases have a relatively rapid progressive symptomatology. The onset may be acute, with paraplegia in a few days. Early diagnosis and prompt treatment to achieve the best possible clinical result, and to keep the patient ambulatory and continent is very important in spinal epidural metastases.

In the Klein et al. study, consisting of pediatric spinal epidural metastases, 30 (17.9%) of 168 patients with Ewing’s sarcoma developed spinal cord compression from epidural tumor spread [12]. In this study, the children’s neurologic function rapidly improved both with chemotherapy and radiotherapy without surgery in small-cell tumors (neuroblastoma, germ-cell, Hodgkin’s lymphoma). On the contrary, patients with Ewing’s sarcoma showed a significant improvement in survival by adding decompressive surgery to treatment. The mean survival was 4.2 years with decompressive laminectomy. In the study of Bacci et al. there were 2 patients with spinal cord compression due to epidural tumor growth, with no radiological evidence of bone involvement [13].

In conclusion, although pure epidural metastases of Ewing’s sarcoma are uncommon, they should be included in the differential diagnosis of an epidural mass lesion. Although local regression is obtained after decompressive surgery and radiotherapy, rapid spread of the disease through the spinal canal and to the cranium, even with subclinical disease, should be considered.

References

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